Gastrointestinal and Hepatic Disease in Systemic Lupus Erythematosus

Brian N. Brewer, MD[a], Diane L. Kamen, MD, MSCR[b,*]

KEYWORDS

- Lupus enteritis • Enteral vasculitis • Intestinal pseudo-obstruction
- Protein-losing enteropathy

KEY POINTS

- Lupus-associated enteritis is a rare feature of systemic lupus erythematosus (SLE) but has morbidity and mortality implications.
- Pathophysiology of lupus enteritis involves a vasculopathic process in the bowel wall with extension to the mesenteric plexus.
- Presenting signs and symptoms of lupus enteritis are often nonspecific but rarely present without other evidence of active SLE.

INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic, potentially severe, frequently disabling autoimmune disease with multiorgan involvement and a typically waxing and waning course. SLE is characterized by the production of a vast array of autoantibodies and is often considered the prototypical autoimmune disease. SLE has the potential to affect virtually every organ, including the gastrointestinal (GI) system, but most commonly presents with musculoskeletal, cutaneous, renal, cardiovascular, hematologic, and/or central nervous system involvement. In contrast with other autoimmune diseases, such as systemic sclerosis and inflammatory bowel disease (IBD), GI system disease activity is rare among patients with SLE. However, for that minority of patients, SLE activity involving the GI system can be severe and even life threatening.

Disclosure: All authors acknowledge that they have no financial or personal relationships with other people or organizations that could inappropriately influence the content of this article. Dr D.L. Kamen is supported by funding from the National Institute of Arthritis and Musculoskeletal and Skin Diseases, NIH under award number K24 AR068406.

[a] Department of Internal Medicine, Augusta University-University of Georgia, 108 Spear Road, Athens, GA 30602, USA; [b] Division of Rheumatology and Immunology, Medical University of South Carolina, 96 Jonathan Lucas Street, Suite 916 e, Charleston, SC 29425, USA
* Corresponding author.
E-mail address: kamend@musc.edu

Rheum Dis Clin N Am 44 (2018) 165–175
https://doi.org/10.1016/j.rdc.2017.09.011
rheumatic.theclinics.com

This article reviews the types of GI system involvement associated with SLE. It includes GI manifestations directly caused by SLE disease activity as well as those occurring in patients with SLE but not directly attributable to active disease. This article is focused on the clinical manifestations of SLE-related GI involvement, with less emphasis on the therapies, which are covered elsewhere in this issue.

WHO IS AT RISK FOR SYSTEMIC LUPUS ERYTHEMATOSUS GASTROINTESTINAL INVOLVEMENT?

The onset of SLE can emerge at any age, but it most often occurs in young women between puberty and menopause. The incidence and severity of SLE is also disproportionately higher among certain racial and ethnic groups, such as people of African descent who live in North America or Europe.[1] In spite of its high impact on individual lives as well as high societal cost, little is known about the cause of SLE.

The clinical symptoms and laboratory manifestations of SLE are extremely diverse. Early diagnosis can be difficult because of the insidious onset of predominantly nonspecific constitutional symptoms (eg, fatigue, joint pains, and low-grade fever). This delay between symptom onset resulting from inflammation, subsequent diagnosis, and initiation of treatment can result in the development of organ system damage. The intent is to identify, when possible, and distinguish between active inflammatory features and features seen in long-standing disease caused by damage.

GI symptoms are common and can occur in approximately half of people with SLE, often triggered by an underlying infection or by medication adverse effects.[2] The most prevalent GI symptoms are nonspecific, such as nausea and vomiting, anorexia, and abdominal pain.[3] Other clinical clues are often needed to distinguish between GI manifestations stemming from an infection, medication side effects, active SLE, and/or a comorbid GI-related medical condition.

DEFINING LUPUS ENTERITIS

By definition, lupus enteritis encompasses a wide spectrum of GI involvement. Lupus enteritis, as part of the British Isles Lupus Assessment Group (BILAG) disease activity index, is defined as either vasculitis or inflammation of the small bowel, with supportive imaging and/or biopsy findings.[4] Abdominal pain is the most common presenting symptom of lupus enteritis, although presenting symptoms vary in character and severity (**Fig. 1**). Studies show a range of 0.2% to 5.8% of patients with SLE affected

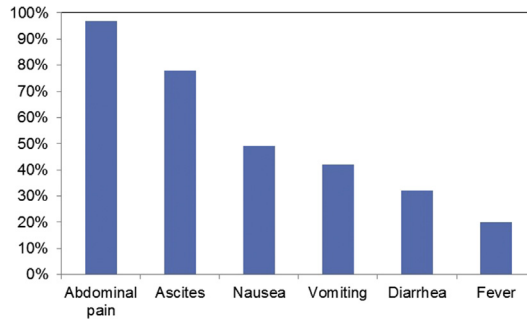

Fig. 1. Incidence of symptoms at presentation with lupus enteritis. (*From* Janssens P, Arnaud L, Galicier L, et al. Lupus enteritis: from clinical findings to therapeutic management. Orphanet J Rare Dis 2013;8:67.)